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Contributions to the Pathology of Infantile Cerebral Palsies.

BY

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By B. SACHS, M. D.,
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About a year ago I had the privilege of reading before the Academy of Medicine a paper prepared in conjunction with Dr. Peterson, and entitled A Study of Cerebral Palsies of Early Life, based upon an Analysis of 140 Cases. If an apology is needed for reverting to the same subject, I offer simply this: that in the former paper we discussed at some length the clinical features of these palsies, and in this article I wish to treat of some of the pathological lesions to be found in these early infantile affections.

It has been my good fortune during the past year to see at least sixty-five additional cases, bringing the total number up to two hundred and five. For this unusual opportunity of studying so many examples of diseases hitherto but poorly understood, I am specially indebted to Dr. Gibney and Dr. Townsend, by whose courtesy the vast

* Read before the New York Neurological Society, April 7, 1891. For illustrations, see Plates I, II, and III.

material of the Hospital for the Ruptured and Crippled has been in part directed to my department at the Polyclinic.

The forms of disease here to be discussed are, after all, somewhat rare, and although peculiar circumstances have enabled me to see more of these cerebral palsies than of anterior poliomyelitis during the past few years, I still believe that the spinal affection is the more frequent; but the large number of cerebral palsies I have seen will give some idea of the relative frequency of these troubles.

In private practice I have seen as many cases of *cerebral* as of *spinal* infantile palsies; this, too, may be either accidental or easily accounted for; but I feel certain that the number of these cases seen by others will increase very considerably as soon as the leading symptoms of these affections are generally recognized. Many of the cases are seen twenty and more years after the onset of the disease; in some, an epilepsy, with very slight hemiparesis; in others, hemiparesis, slight contractures, posthemiplegic movements, and exaggeration of the reflexes are the only symptoms that point to a cerebral affection of old standing. I have seen at least six cases of epilepsy which have passed through able hands and have been pronounced cases of typical epilepsy, but which on closer examination have been found to be instances of epilepsy associated with early spastic cerebral palsy, both conditions being due to one and the same lesion.

Large orthopædic institutions, pauper asylums, and homes for idiots are full of these patients. In an able statistical paper on club-foot, published in the *Transactions of the Medical Society of the State of New York* for 1890, Dr. Townsend collected seven hundred and twenty cases due to spinal lesion, and seventy-seven spastic cases (say one to ten), mostly due to cerebral lesions; but Dr. Townsend has been

kind enough to tabulate for me the cases seen at the Hospital for the Ruptured and Crippled during the last year, and, with the full knowledge of the subject now at his command, he finds the relative frequency of cerebral cases far greater than any one would have expected it to be.

The records of the hospital show for the past year one hundred and forty-two spinal cases, with deformities of all sorts, and ninety-one of cerebral origin. Doubtful cases were omitted from the list. These figures, which are larger than any other institution could show, prove that, of the cases seen during one year, 61 per cent. were of spinal origin and 39 per cent. of cerebral; the proportion is not three to one. Dr. Townsend, in a letter to me, expresses his great surprise. I should not be surprised if the proportion should be found to be still greater in favor of cerebral palsies.

The clinical features of these cerebral palsies were so fully given in the paper* alluded to that I have to-day but to add that the symptomatology as established by the former studies has been fully borne out by an analysis of cases seen since the publication of that paper.

Let me simply remind you of a few of the points which we sought specially to fix upon. First, that the mere form of the palsy, while pointing to a different location and a varying extent of the lesions in question, should not constitute a sufficient reason for the creation of special types, and that a hemiplegia, a diplegia, or a paraplegia may be due to the same morbid processes. Secondly, that although the majority of the cases are of the distinctly spastic type, flaccid paralyses do sometimes occur which are unquestionably of cerebral origin; and, again, that there are a few atrophic palsies due to lesions in the brain and not to lesions in the spinal cord or the peripheral nerves. These features, taken in conjunction with the occurrence of dementia or

* *Jour. of Nerv. and Ment. Dis.*, 1890.

epilepsy and with the typical contractures and exaggerated reflexes, make the diagnosis a very positive one, and I can safely say that of the two hundred and five cases we have seen, doubt as to the spinal or cerebral origin was entertained in but a single instance.*

Accepting these clinical features, we are ready to pass on to a consideration of the pathological lesions underlying them. The most important conclusion of former studies on this point was that the largest majority of these cerebral palsies of childhood were due to hæmorrhage, thrombosis, and embolism. This has been borne out, not only by the publications of others, but by my own recent studies. I have found a confirmation of my views in post-mortem examinations, and, much to my gratification, in a recent operative case with Dr. Wyeth, in which the diagnosis of epilepsy and hemiparesis due to meningeal hæmorrhage thirty years ago was corroborated as soon as the motor arm center in the right hemisphere was exposed.

Not wishing to weary you with the details of these pathological studies, I have prepared a table in which I make the attempt to give the morbid lesions and the symptoms which most frequently accompany them. Like every such attempt, this one will be imperfect; but I trust that additions and corrections to be made hereafter, by others if not by myself, will make good whatever defects there may be in the classification submitted to you. In this table I have divided the cases into those of truly congenital origin, those occurring during birth, and those that may be properly called acute or acquired cerebral palsies.

* Since the foregoing was written, I have seen one case, and the only one, in which there was evidence of a spinal *and* a cerebral lesion.

	Morbid lesion.	Form of palsy.*	Distinguishing symptoms and conditions.
PRENATAL.	Large cerebral defects.	Diplegia, hemiplegia, paraplegia.	Birth normal, possibly premature; convulsions often absent; mental condition sometimes good, often deficient.
	Agenesis corticalis and minor lesions. (Highest nerve elements involved.)	Diplegia, hemiplegia, paraplegia.	Birth normal; paralysis may be flaccid; great mental defect; convulsions present or absent.
BIRTH PALSIES.	Meningeal hæmorrhage, rarely intracerebral hæmorrhage (later conditions: meningo-encephalitis chronica, sclerosis, and cysts).	Hemiplegia, diplegia, paraplegia.	Protracted labor, chiefly in primiparæ; instrumental delivery; asphyxia at birth. Convulsions soon after birth and oft-repeated; early development of contractures. Idiocy frequent.
ACUTE (acquired) PALSIES.	Hæmorrhage (meningeal and rarely intracerebral), thrombosis (sometimes due to syphilitic endarteritis). Embolism. Later conditions: atrophy, cysts, sclerosis, and softening.	Hemiplegia, diplegia, paraplegia.	Sudden onset, generally with convulsions; repeated convulsions; slight or no fever; recovery of leg or arm very frequent; post-hemiplegic movements. Mental development rarely interfered with; after or during acute infectious diseases.
	Meningitis chronica.	Hemiplegia, diplegia?	History of onset after attack of meningitis. Basilar symptoms with spastic paralysis; basilar symptoms sometimes wanting.
	Hydrocephalus (rare as sole cause of palsy).	Paraplegia, diplegia, hemiplegia.	Increasing size of head; onset of paralysis gradual; progressive deterioration of mind; exclusion of other lesions.
	Primary encephalitis? (Strümpell.)	Hemiplegia, diplegia.	Onset with fever and convulsions, independently of or after acute infectious diseases.

* The form of palsy is given in the probable order of frequency.

I feel compelled to explain a few points. First, that hæmorrhage, thrombosis, and embolism were positively proved to be the most frequent lesions in the acute cerebral cases; that other conditions so often cited were terminal and not initial morbid states. It may be surprising, too, to many, that primary encephalitis is put at the very end of the table, and with a question-mark at that. Strümpell's now famous theory has taken a wonderful hold upon the medical mind, and yet I must repeat what I said last year—that there is remarkably little proof of the existence of this condition. The reasons for this statement must be given before proceeding to a discussion of the true morbid lesions. Reasoning by analogy, Strümpell concluded that a certain number of cases of acute infantile cerebral palsies were similar in every respect to cases of poliomyelitis anterior, except that the symptoms pointed distinctly to a cerebral and not a spinal trouble. It was natural, therefore, for him to maintain that the gray matter of the cortex was subject to the same changes that affect the gray matter of the spinal cord; hence, if we know of a condition of poliomyelitis anterior, why should there not be a condition of polio-encephalitis? There is no inherent reason why not; but proof is wanting. Strümpell himself has abandoned the idea of a polio-encephalitis, and now speaks of primary encephalitis; but even this theory must be accepted with some reserve. The more fascinating the idea, the more carefully we should examine into it. The only proof we have of the probable existence of this disease in children is given by two well-observed cases of Möbius,* in which one child was affected by a poliomyelitis and another child in the same family at the same time developed typical acute cerebral palsy (spastic hemiplegia without aphasia). Furthermore, Strümpell †

* Schmidt's *Jahrbücher*, 1884, cciv, p. 135.

† *Dtsch. Arch. f. klin. Med.*, xlvii.

has reported within the last year two cases of adult apoplexy in which he was surprised to find a widespread encephalitis and not hæmorrhage or embolic softening, as every one would have been apt to believe.

I can not allow that the occurrence of such a condition in the adult is sufficient proof of its occurrence in the young; but even granting the analogy, all that we can infer is that among the many thousands of cases of adult apoplexy a very small number may be due to an encephalitis, and it is possible that an equally small or smaller number of these infantile cases may be due to the same condition; but all the evidence we have is entirely against the assumption that a primary encephalitis is the rule in the acute brain palsies of children. I uphold what I said a year ago—that polio-encephalitis should be diagnosticated last, not first, and I can not understand why this encephalitis should always take the form of a hemiplegia, as Strümpell would have it. I have seen at least two cases in which, if in any, the symptoms of onset corresponded accurately to the description given by Strümpell, but the form of paralysis was a diplegia and not a hemiplegia. However much one may be inclined to give this theory its due weight, it has undoubtedly been a hindrance to the understanding of the pathology of the diseases in question. We might as well allege cysticercus to be the most frequent form of brain tumor in children as to maintain that a primary encephalitis is the most frequent cause of spastic palsy in children, and to raise these rare and doubtful cases to the dignity of *the* cerebral palsy is an absurdity.

From the tables which I have submitted it is evident that there are many morbid states giving rise to the same symptoms. Of the three autopsies which I have had, each one represented a condition entirely different from the other two. Two of these three autopsies I wish to explain to-

night.* The first case is not only interesting as a contribution to the pathology of this special subject, but would be able to stand on its own merits if reported, for the tumor developed in the brain shortly before death. The history of this first case is as follows:

J. W., one of the one hundred and forty patients, a healthy-looking boy, eight years of age, was well until six years and a half old; then without any known cause was seized with convulsions and developed right hemiplegia. When brought to the Polyclinic, three months later, he presented the symptoms of a typical spastic hemiplegia of the right side, including the face, and in this permanent involvement of the face presented at least one unusual feature. He furthermore gave the history of loss of speech following the attack, and of repeated epileptic seizures involving the right hand only. His mental development was only a little retarded, while his head showed the hydrocephalic condition, and during observation the head was noticed to increase in size. No other deformities of the skull. It was a mere matter of chance that this case sought admission to the Montefiore Home, where he continued under my observation. While there he was doing extremely well, was happy, ran around freely, and there was every prospect of his remaining an inmate for many years. No changes were noted until the end of October, 1890, when one day he suddenly fell down and the right hemiparesis of old standing had developed into complete hemiplegia. There was no loss of consciousness during this occurrence. The face was not more paralyzed than before; there was no incontinence of urine and fæces. Within a few days after this fall the boy developed fever varying between 101° and 103·5° F. During this time a disturbance of vision was developed which ended in absolute blindness. Speech became more and more difficult; he developed a mild stupor which gradually deepened into coma. Two weeks later intermittent

* For an account of the third case, see *Jour. of Nerv. and Ment. Dis.*, Sept. and Oct., 1887.

opisthotonos was observed, this condition, too, becoming permanent. After five weeks, with the continuance of all the other symptoms, left ptosis and paralysis of the left rectus externus and anæsthesia of the right cornea were added; the pupillary reflexes were lost, and the contractures of the right side increased. Dr. Koller, who made an ophthalmoscopic examination, reported that the papillæ were swollen and whitish-gray, not distinctly outlined, arteries very small and thread-like, veins dilated and tortuous—in short, a retrogressive papillitis. From the time of the fall until deep coma had been established the slightest touch over the left motor area was extremely painful, the contractures became extreme, the reflexes were exaggerated throughout, the anæsthesia of the cornea became absolute, and for fully thirty-six hours before death the right eye ceased winking; respiration became irregular, and the boy finally died eight weeks after the onset of this last attack. Convulsions were not repeated during the whole of this time.

The old hemiplegia was ascribed to a cystic formation probably due to hæmorrhage over the left motor area. For a few days after the development of the increased paralysis and during the continuance of the fever the idea of abscess in connection with the old cyst was entertained; but this was soon abandoned, and a tumor growing in the walls of the old cyst was the diagnosis made in the presence of the house staff and adhered to to the end. In no other way could the increase of the former hemiplegia, without the addition of further symptoms, be explained.

The tumor was supposed to be a glioma or gliosarcoma, but the development of basilar symptoms later on made me hesitate and led me to believe that a basilar meningitis had been developed, and it was therefore natural to suppose that the original tumor, although the boy was in excellent health, was of a tubercular nature. In this I was mistaken, for the basilar symptoms must be attributed directly or indirectly to the second tumor found in the right temporo-sphenoidal lobe.

The autopsy was performed within twenty-four hours after death. The skull and the spinal canal were the only parts opened. The skull was large and thin, with enormous occipital

bulging. The fontanelles were closed. The calvaria was easily removed, there being no adhesion between the dura and the skull. The unusual size of the brain was at once made evident, the posterior portion having been enormously expanded by the accumulation of fluid within the ventricles, and the hemispheres being so thin that the fluctuating mass beneath could be distinctly felt and all trace of normal fissuration was entirely lost. The conditions are best explained by reference to Plate I, Plate II, and Fig. 1 in Plate III.* While the brain was still *in situ*, the cyst occupying the left motor area and the tumor in its walls could be distinctly observed. After the brain had been removed, further changes were observed at the base; first of all a large cystic formation† at the apex of the left temporo-sphenoidal lobe, pressing in upon and almost covering the left crus. The optic nerves were evidently atrophied, and the left abducens was extremely thin. There was no thickening of the meninges in the interpeduncular space nor over any part of the base. Pushing the basilar cyst aside, the degenerated condition of the left crus was extremely marked. Next in order we came upon the large and hard tumor in the right temporo-sphenoidal lobe. Upon puncturing the anterior cyst, large quantities of fluid escaped—surely more than a pint—and with it the posterior bulging diminished, showing that this anterior cyst had communicated with the ventricle. A small portion of the tumor in the right temporo-sphenoidal lobe was at once removed for examination, but with this exception the brain was submerged *in toto* in Müller's fluid, in which it was allowed to harden for a few weeks before any further examination was made. The spinal cord was removed and suspended in Müller's fluid. The brain and spinal cord have been made the subject of careful examination, which has yielded the following facts:

The unusual weight of the brain was caused by the accumulation of fluid in the ventricles, and the two tumors. The right

* Special acknowledgment should be made to Dr. Macdonald for the beauty of his drawings and his careful study of the specimens submitted to him.

† The cyst, in reality, formed part of the apex of the temporo-sphenoidal lobe.

hemisphere was slightly œdematous, very much flattened in its occipital portion, but otherwise normal; the pia could be removed freely from every part of its surface. The left hemisphere appeared larger than the right, in consequence of the greater amount of fluid in the left ventricle. All the blood-vessels were of normal appearance. The pia could easily be removed, except over the cyst; here no pia could be made out; it was evidently an integral part of the roof of this cyst.

The old cyst (Plate II) occupied an area bounded anteriorly by the post-central convolution, posteriorly by the anterior portion of the interparietal fissure, while it extended upward within an inch of the chief longitudinal fissure, and downward as far as the upper end of the fissure of Sylvius. It had thus involved a portion of the posterior central convolution, the superior and inferior parietal lobules, and the gyrus supramarginalis. The roof of the cyst was formed by a thickened, somewhat transparent, membrane. On cutting through this membrane, it was evident that there was a very large amount of detritus in the cavity of the cyst, while in the lower anterior end of this cavity a hard, small, round tumor is felt. Under the surface the destruction has evidently exceeded these limits, and the whole of the posterior central convolution was undermined and softened. The posterior portion of the left hemisphere in the hardened specimen, though now very much contracted, offers far less resistance to pressure than the portion frontad of the sulcus Rolando does.

The actual size of the "small round tumor" in the cyst was not suspected until vertical sections were made through the brain many weeks after it had hardened in a bichromate solution. You will all grant that the term "small round tumor" does not do justice to the facts (Plate II, Fig. 2). The part protruding in the cyst was a minimal portion of this huge tumor. As you see, it extends to the ventricles from the outer margin of the cortex, pushing everything aside, and leaving only here and there some normal cortical tissue. It begins, furthermore, at the anterior end of the Sylvian fissure and extends well back to the beginning of the occipital lobe; it has entirely displaced the ventricle, which would be difficult to recognize were it not

for the great thickness of its ependyma. Sections of this tumor, stained in carmin, picrocarmin, and hæmatoxylin, prove it to be a small-celled sarcoma in which there are also some spindle-shaped cells surrounded by a large amount of fibrous tissue.

The base presents a larger number of pathological changes. In the hardened specimen we are struck first of all by the collapsed condition of that portion of the left hemisphere lying caudad of the fissure of Sylvius. Next we observe the peculiar cystic condition of the inner part of the temporo-sphenoidal lobe which pressed seriously upon the left crus. In the right temporo-sphenoidal lobe, about two inches below its apex, a large tumor was found extending from the outer surface as far inward and downward as the ventricle, and this tumor has all the characteristics of the tumor in the old cyst, is uniformly hard to the touch, and, as the microscopical examination has shown, is also a small-celled sarcoma. Both olfactory nerves are extremely thin; the optic chiasm and optic tracts had been pushed aside and are distorted in consequence of the growth of the cyst of the left side (Plate III, Fig. 1). The left optic tract is very much smaller than the right. The left crus shows such marked atrophy of the middle portion that the internal and external portions almost fall upon themselves. The right crus is broad and normal. The left third nerve is very much compressed; the right third appears normal. In the fourth and fifth nerves no changes are to be observed, nor in the macroscopical appearances of any of the other cranial nerves. The pons shows an inequality in favor of the right side, while in the medulla this is not so distinct.*

The microscopical examination of the brain and cord showed changes which were in part visible in the fresh specimen, namely, very marked secondary degenerations in the left pyramidal tract of crus, pons, and medulla, and throughout the entire lateral columns of the right side of the cord. There is also a slight degenerated area in the left lateral column. Changes in the anterior columns are not very distinct.

* In Plate III, Fig. 1, after the hardened specimen, some of these points are not so distinct as they were in the fresh specimen.

In spite of the manifold lesions found in the brain of this child, there is no difficulty in explaining the relations between these lesions and the symptoms during life, nor in determining the sequence in which these various pathological conditions became established. That the old hemiplegia was due to the cyst in the left hemisphere there can be no doubt. Some doubt there may be, however, as to the origin of this cyst, but, in view of the peculiar detritus, of the very irregular margin of the cyst, we need have no hesitation in assuming the hæmorrhagic origin of this formation. Moreover, the area involved in the cyst does not correspond to the territories supplied by the main branches of the middle cerebral artery; there is no reason, therefore, to think of an initial embolic softening, and, more than that, the blood-vessels were found normal. The existence of the hydrocephalus probably antedated the cyst in the left hemisphere. It was specially noted in our records that the head was increasing, and I have no doubt that the smaller cystic formation near the apex of the temporo-sphenoidal lobe was of very recent development. The other changes at the base were the result of compression or of the secondary degeneration following upon the destruction of a large portion of the left motor area.

We have learned in the course of our studies not to attach too great an importance to the hydrocephalic condition pure and simple. There are very few cases in which the hydrocephalus was the cause of the infantile cerebral palsy. In an analysis of one hundred and four cases with autopsies, not a single case could be attributed to this condition, although other lesions were often found associated with a hydrocephalus. Inasmuch, too, as our little patient was bright and well until eight weeks before his death, we can assume with perfect safety that, however much the hydrocephalus may complicate the anatomical appear-

ance of the brain, it was not responsible for any of the symptoms.

It is not a little surprising to find that the cortex will tolerate great hydrocephalic pressure and yet not suffer in its functions. I have no doubt that the occipital cortex in this case performed all its functions normally until a few weeks before death. The tumor unquestionably increased the hydrocephalic condition, but the head was noted to be large and increasing long before the symptoms of tumor appeared. And remember that in this, as in so many other cases, the general intelligence remained good. I was curious to compare the cortical structure of the occipital region with those of other regions less affected by the hydrocephalus; pieces of cortex were therefore excised from the right and left occipital regions and from the left frontal region.

The examination of these specimens showed, first of all, the marked diminution of gray matter in the parts much compressed. In the three specimens which I present,* you will observe that in the one representing the extreme hydrocephalic condition there is scarcely a normal pyramidal cell to be detected; the blood-vessels are small, and some of them show a slight cellular proliferation of the vessel walls; in the other specimens these conditions are less marked, and in the frontal section, where the hydrocephalus exerted the least influence, you find a tolerably normal condition of the cells, but a surprising dilatation of the veins, and many of these veins are completely filled with blood. But in all these sections there is no evidence of sclerosis.

I was so firmly convinced of the existence of a hæmorrhagic cyst in this case of acute cerebral palsy that I was at once ready to explain the lesion which could give rise to an increase of the old symptoms. Some new process, I

* The demonstrations were made at the meeting.

argued, must have started up in the old lesion, and the walls of the cyst seemed to me to offer the very best opportunity for the incursion of a growth. The diagnosis of such a tumor was made easy by the rapid development of double optic neuritis, of blindness, and, above all, by extreme painfulness over the left motor area. Considering the well-known fact that tumors are most apt to develop in atrophic or mal-developed tissues, and considering, furthermore, the relative frequency of these cases of cysts in the brain, it is surprising that so few instances of tumor development in old cysts have been reported. I have carefully examined the writings of Bernhardt,* of Nothnagel, † of Mills, ‡ and of Starr # bearing upon this subject, and have not found the records of a single case exactly like my own. Cysts of all dimensions are frequent enough, and in three cases only—those of Jackson, ‖ Moines, △ and Escribano ◊—is there any note of the joint occurrence of cyst and tumor; but in none of these cases, even, was there any good reason to believe that the cyst was developed in childhood, or that it antedated by any considerable length of time the development of the tumor. Microscopic examination has shown that the two tumors in this case are of exactly the same character, although the one at the base is much smaller than the one in the left hemisphere. There is every reason to believe that it was developed later than the one which caused the hemiplegia. The tumor at the base, however, is evidently responsible for the symptoms which were developed toward the end of life, which pointed to an in-

* *Hirngeschwülste*, Berlin, 1881.

† *Topisch. Diagnost.*, etc., 1879.

‡ Pepper's *System of Med.*, v.

Keating's *Cyclop. of Dis. of Children*, iv.

‖ *Med. Times and Gaz.*, Nov., Dec., 1872.

△ *Arch. f. path. Anat. u. Phys. u. f. klin. Med.*, lxx, 1877. Schmidt's *Jahrbücher*, 1867, i.

volvement of many of the cranial nerves, and which led me on this very account to suspect that there was a basilar meningitis.

In view of the unusual size of the tumor, I wish to allay any doubts as to its relation to the old hemiplegia. If the tumor was the cause of the old hemiplegia, what were the symptoms of the cyst? Surely not the increase in the hemiplegia, for the complete disintegration of the contents of the cyst proved them to be far older than two or even six months. I can not positively assert that the tumor began to develop a few months before death; it probably began a little earlier, and had evidently attained a respectable size before it produced prominent symptoms; but the sudden accession of headaches, double optic neuritis, and semi-coma point to a sudden and rapid growth. That there were no local convulsions need not surprise us if we remember that the old cyst left very little motor cortex capable of "discharging." The size of the tumor, astonishing as it is, is not an indication of great age, for sarcomata are known to grow with extreme rapidity in the brain and in other organs. I remind you of the sarcomata, many pounds in weight, that have developed within a few months in the breasts of young women. In this instance we are again reminded of the fact that it is quite impossible to foretell anything regarding the size of a brain tumor. The tumor in this case, though the most striking morbid condition, need not detract from the importance of the old cyst, which I hold to be the residue of a subpial hæmorrhage three years ago, the cause of this acute form of infantile cerebral palsy.

The second case which I shall take the liberty of reporting is of a very different character from the first—different in its clinical symptoms and in the pathological findings. As this case is to be reported in full, together with a series of studies by Dr. Holt and myself, I shall give

but brief details this evening. The patient's history was given in the paper of last year, from which I quote the following:

The child was a little boy one year of age. He had congenital spastic paraplegia. The mother, a primipara, was in good health, but the labor was hard and dry for forty-eight hours. The child was asphyxiated when born. From the first day up to the age of six months and a half the child passed through a rapid succession of tonic and clonic spasms affecting all the muscles of the body, causing rigidity of all extremities, opisthotonos with extreme arching of the back, enormous exaggeration of all the reflexes, including a patellar clonus on the slightest excitation. There was convergent strabismus and continual crying. The mental condition, as far as could be judged, was deficient but not absolutely idiotic. The epileptic spasms were so frequent that we succeeded in obtaining a photograph of the child during one of these spasms. In the intervals between the spasms the child could move its hands and arms, but the legs remained in the condition of spastic rigidity. As the parents confessed themselves unable properly to care for the child, we referred it to Dr. Holt's Babies' Hospital, where the child remained for a few weeks, passed through an acute eruptive disease of doubtful character, and finally died. The autopsy was made within twenty-four hours after death by Dr. Holt, who has kindly left the brain to me for detailed examination. The most noticeable feature of the brain was the absolute adherence of the pia over both hemispheres: it was impossible to remove even the smallest strip from any part of the cortex. The brain appeared somewhat firmer to the touch, but, as far as the fissuration could be determined through the adherent pia, there was no unusual change in this respect. But most striking of all was the marked symmetrical atrophy of the frontal halves of both hemispheres. The brain and cord have been carefully hardened in Müller's fluid, and the cortex, the ganglia, and the whole spinal cord have been made the subject of careful microscopical examination. The change in the cortex is very evident on specimens stained in carmine, picrocarmin, and hæmatoxylin.

From whatever part of the cortex specimens were examined, the same change was shown throughout. We have, first of all, a general adhesion of the pia to the cortical tissue; the pia is thickened and characterized by a general cellular infiltration; its blood-vessels show marked cellular proliferation; the veins in the subpial space are dilated and filled with blood, and small branches of the pial sheath passing in between the convolutions show the same changes that we find in the pia that is stretched over the convex surface. In the cortex itself, instead of the normal arrangement of cells, we find few if any normal pyramidal cells; but in the outer layers, and particularly in what would correspond to Meynert's third and fourth layer, we find an enormous profusion of small glia cells. The blood-vessels are in part normal, but many of them show marked small-cell proliferation of their walls. There is unquestionably a thickening of the neuroglia. The anatomical diagnosis is clearly that of a chronic meningo-encephalitis. In the drawing (Plate III, Fig. 2) the space *a* is somewhat artificial. The changes in the cells and in the vessel walls are distinctly shown; to the right of the pial projection there is a structureless area, probably the result of a small hæmorrhage. We can not err in attributing this meningo-encephalitis to a very wide-spread effusion of blood between the pia and the cortex at the time of birth. Protracted labor and the marked asphyxia are the clinical conditions which corroborate that view. The spinal cord, as far as it has been examined up to the present time, reveals a most distinct degeneration of both lateral columns. That this degeneration is secondary to the cortical lesion there is every reason to believe, for this degeneration can be recognized in all parts of both motor tracts and in the pyramidal tracts of the pons and medulla, as well as in the lateral

columns at every level of the cord. It must be remembered that the cortical affection was bilateral.

I have only to add with reference to this case that it has a special significance, inasmuch as it is the second case of congenital spastic paraplegia in which an autopsy has been made, or at least recorded; and it is of extreme value, it seems to me, as proving, beyond the shadow of a doubt, that these cases of congenital spastic paraplegia are of cerebral origin and not due to spinal-cord lesions. In the only other case of infantile spastic paraplegia dating from birth which has been recorded with autopsy, Förster* found a general sclerosis of the brain; this may have been a secondary condition, but the case loses much of its value since no microscopical examination was made. Only within the last few years so eminent an observer as Ross† was inclined to think these cases of spinal origin and due to traction at birth. I have elsewhere stated that the occurrence of idiocy and mental enfeeblement in fully 80 per cent. of the cases of congenital spastic paraplegia would of itself seem to point to the brain as the source of the disease; and if, as in my own case, the lateral columns are affected, the affection is secondary to the initial cerebral lesion. The unusual convulsions in my case must be attributed to the fact that there was an active and widespread encephalitic condition; this inflammatory state was not at an end before life terminated, and the constant occurrence of epileptoid seizures is what we might expect in such a condition as the one I have just described.

The knowledge of the pathological lesions underlying these various conditions is not only a matter of scientific curiosity, but it has a most distinct bearing upon the

* *Jahrb. f. Kinderheilk.*, xv.

† On the Spasmodic Paralyses of Infancy. *Brain*, v.

treatment of these diseases. Hitherto the majority of cases have either not been treated at all or else they have helped to swell the numbers in orthopædic and other hospitals, and many of them are the inmates of our pauper institutions. The orthopædic surgeon will have to attend to the correction of the physical deformities; in those cases in which a persistent palsy, or more especially an extreme contracture of one or both of the extremities, is the chief residue of the disease, the surgeon will no doubt sooner or later be able to give a fair measure of relief. But it is more important for us to consider whether we have a right to interfere with the cerebral lesion in order that we may combat the development of idiocy or epilepsy, or whether it be possible—one or the other of these conditions having been established—to obtain relief by surgical procedures. The question can not be decided unless some such analysis of the morbid lesions be attempted as I have given in this paper. If we have to deal with a condition of porencephalus or with a condition of cortical agenesis, it is manifestly impossible to improve this condition by an operation; and if such brains are incased within a microcephalic skull, even the recent operation of craniectomy will do no earthly good. As for the birth palsies, no surgeon will be bold enough to attempt the removal of a subpial or subdural clot in a child only a few days old; and long before the child is strong enough to tolerate any such serious operation, the mischief is done, an encephalitic or a sclerotic condition has been established for which the surgeon's knife can promise no relief. It is only in the cases of acquired cerebral palsies that one would naturally think of the possibility of operative procedure. Should the diagnosis point to thrombosis or embolism, there is no good reason for surgical interference. But how about cases of

hæmorrhage?* Arguing from the analogy with adult cases, there would seem to be no good reason to interfere in cases of hæmorrhage until some time has elapsed after the initial apoplexy; for presumably, as in adult apoplexy so also in the infantile forms, much of the clot will be absorbed; but if we leave the clot to be absorbed, a very short time may suffice for the establishment of secondary changes in the brain and spinal cord. In adults the clot is generally in or near the ganglia; in children it is most frequently upon the surface. It is my belief that the greatest good will be done if the surgeon will exercise his skill at this early period.

I have no hesitation, therefore, in recommending that, if the symptoms point to the formation of a large clot over the motor area of a child otherwise healthy and strong enough to endure the operation, the attempt be made to remove the clot in order at once to release pressure upon the given area and to prevent secondary degeneration. It is commonly urged that in many cases we can not at the start state with any degree of certainty whether the case is a spinal or cerebral affection. This I can not concede except in the rarest cases. In several instances I have made the differential diagnosis at once, and in others within twenty four hours after the onset of the trouble, and I claim no special diagnostic powers. The excellent recoveries from surgical procedures upon the brain have led up to this view, and I firmly believe that recent cases will give more satisfactory therapeutic results than the cases of old standing. My own experience covers three cases of infantile spastic palsies of old standing in which the patients have recently been operated upon—one by Dr. Gerster and two by Dr. Wyeth. The time is too short to speak of the ultimate result as regards the persistence of the epilepsy, and,

* It goes without saying that this diagnosis is sometimes impossible.

of course, we do not expect the surgical operation upon the brain to improve an old-timed hemiplegia. In one of the three cases, as I intimated at the beginning of this paper, a hæmorrhagic infiltration and discoloration of the pia, with marked thickening of the same, was found as proof of the old meningeal hæmorrhage. Small vertical incisions were made through the pia at this point by Dr. Wyeth to release the local pressure. In the other two cases, although the dura was opened, no change could be made out. There are several possibilities which must be considered:

First, every trace of the old trouble may have disappeared; and, secondly, in cases of meningo-encephalitis or of sclerosis the changes can only be made out by microscopical examination. Take the case of Frobel, the child with spastic paraplegia, and you will readily perceive that if its brain had been exposed *intra vitam*, the changes in it might not have been recognized. Next we must consider whether, if we have good reason to suspect the existence of a cyst or of a scar, it would be wise to make this the point of attack. From the fact that a cyst acts somewhat like a foreign body in the brain, there could be no harm in removing it, or at least in emptying its contents, provided drainage of this cyst does not mean withdrawal of a large portion of the cerebro-spinal fluid, and many cysts are entirely independent of other parts and cavities of the brain. But the operation would surely do no good if done as late in life as in Dr. William A. Hammond's case, in which the patient, who was twenty years of age, had carried the large cyst from the very earliest period of life.* The presence of a cyst unquestionably acts as an irritant upon the neighboring brain area; it is therefore in cases of persistent hemi-epilepsy associated with infantile hemiplegia that the attempt to remove the cyst or other morbid lesion might be attempted

* *N. Y. Med. Jour.*, 1890.

justly enough. Cases of diplegia and paraplegia associated with epilepsy do not appear to be the proper cases for operation, for to do any reasonable amount of good, both halves of the brain would have to be operated upon. This should not be attempted unless operations upon cases of hemiplegia with epilepsy have shown that some good can come from such an operation.

The matter becomes still more difficult in those cases in which a general lobar sclerosis or a condition of primary encephalitis is suspected. In lobar sclerosis the disease is always widespread, and if one area was removed, the remaining ones would be sufficient to continue many of the symptoms. Meningo-encephalitis has an acute and a chronic state. In the acute state we would surely not operate, and in the chronic state we can not say exactly how far the inflammatory process has progressed, and we could do no good unless after removal of an encephalitic area the child's brain was sufficiently young to take upon itself functions that were lost. And let us reflect that if this could occur in a given case, we should have a right to expect that the injured brain before operation would assume the functions of parts that through disease were practically lost to it.

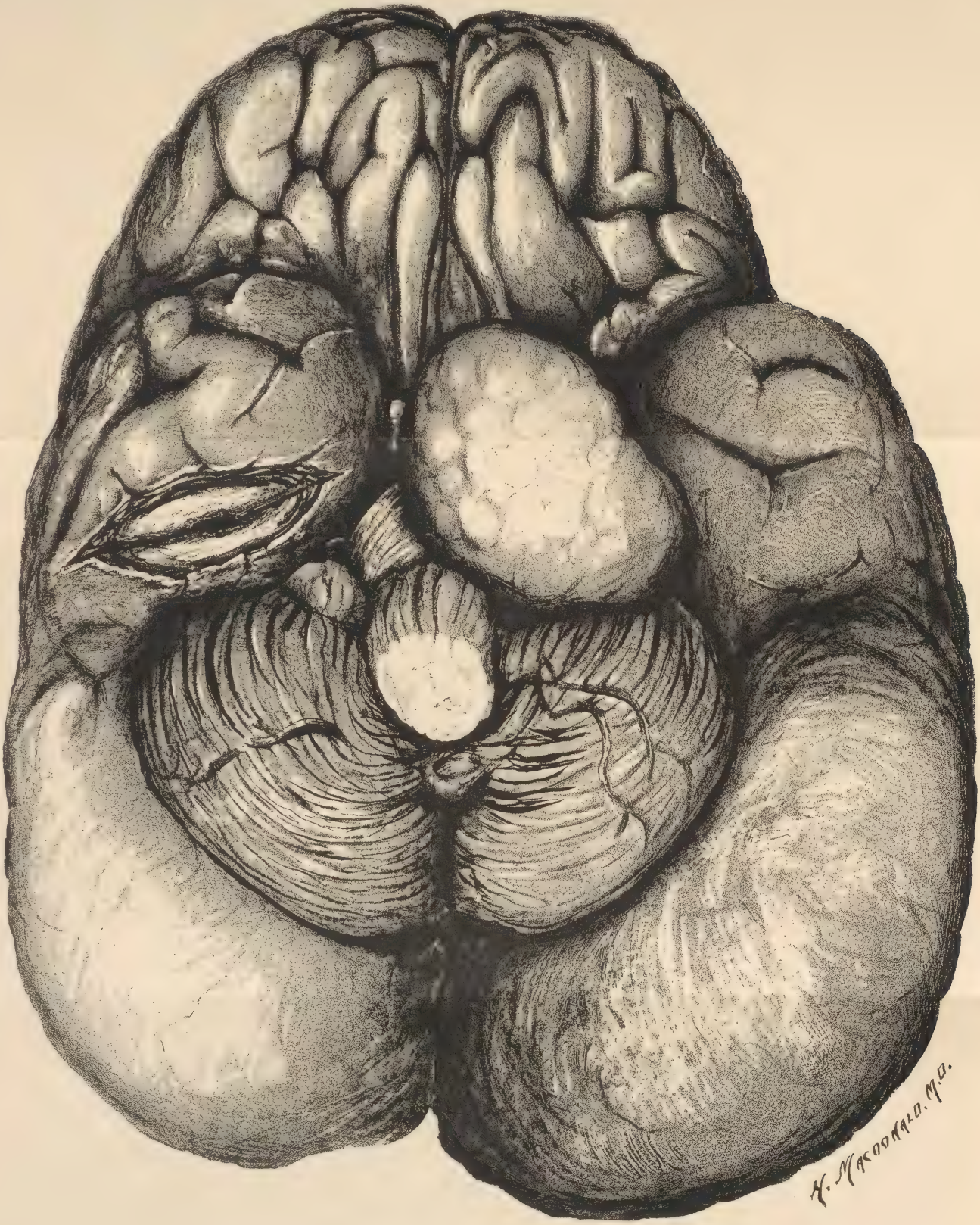
There are two facts, moreover, which are well calculated to dampen our surgical ardor: First, a brain that has been the subject of epileptic discharges falls into what one may justly claim the epileptic habit. Remove the original focus of disease, and the habit or other foci of disease may remain. My own experience in the surgical treatment of focal epilepsy leads to anything but a roseate view. Secondly, granting that the removal of a clot or an old infiltration or a scar may inhibit the epilepsy, you may and probably will have to face the alternative of a palsied arm or leg or both. Considering how useless many of these con-

tractured, athetoid, and paralyzed extremities are, the choice would be an easy one for us; but it is eminently proper that the patient, if old enough, should express his preference.

And yet there is promise enough among this special class of cases, and if we are to fix upon the one of a dozen cases of infantile palsies in which the surgeon may come to the rescue, we must, above all, have a thorough understanding of the morbid lesions.

PLATE I.

DRAWN FROM THE SPECIMENS BY DR. H. MACDONALD, AND REPRODUCED DIRECT.



Ventral aspect of brain, showing bilateral hydrocephalus, cysts, and tumor.

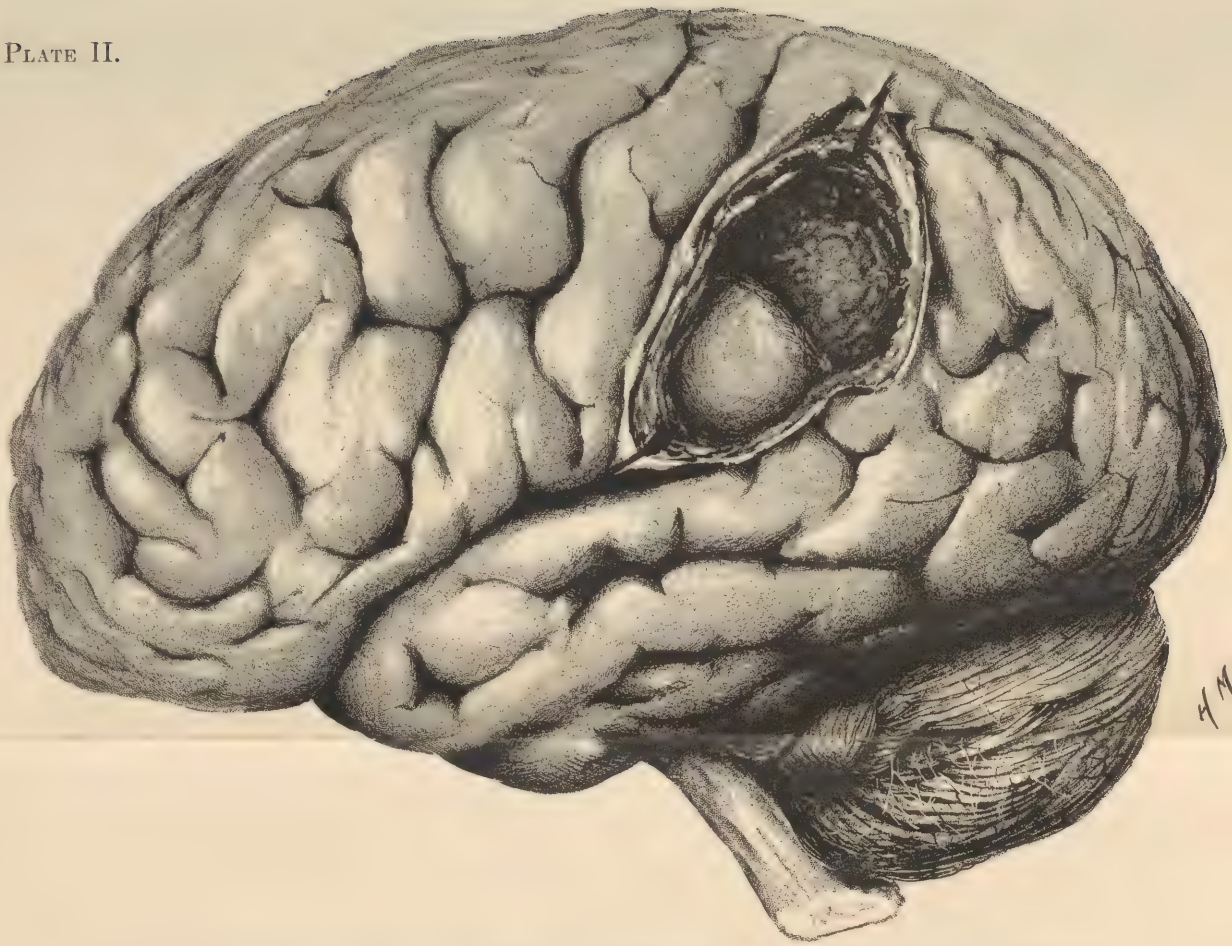


Fig. 1.—Showing position of cyst with portion of tumor. Roof of cyst has been laid open by dissection. The hydrocephalic enlargement is omitted to economize space.

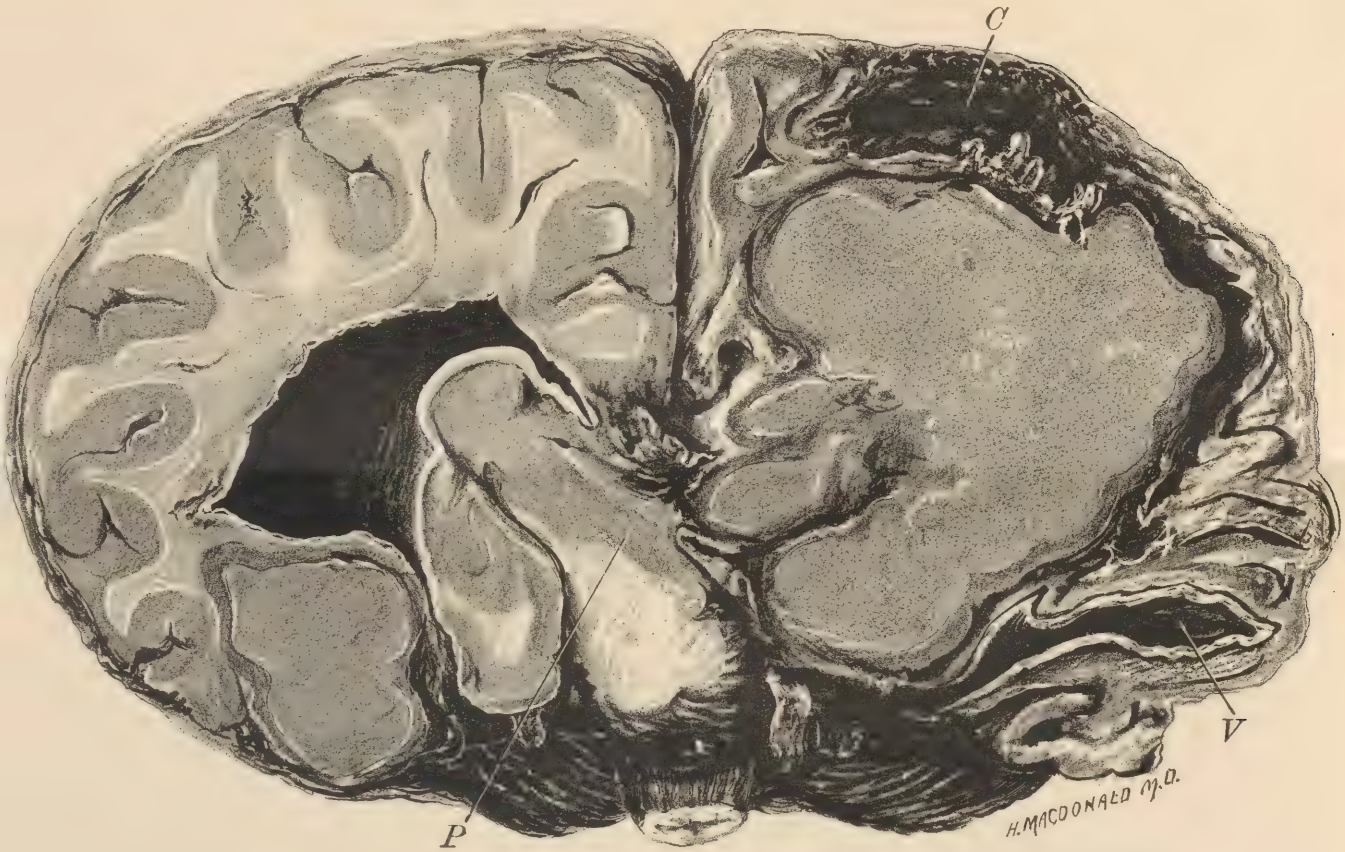


Fig. 2.—Vertical section through cyst and both tumors, showing also distortion of the brain axis and displacement of the left ventricle. *C*, the cyst. *P*, the pons. *V*, the left ventricle.

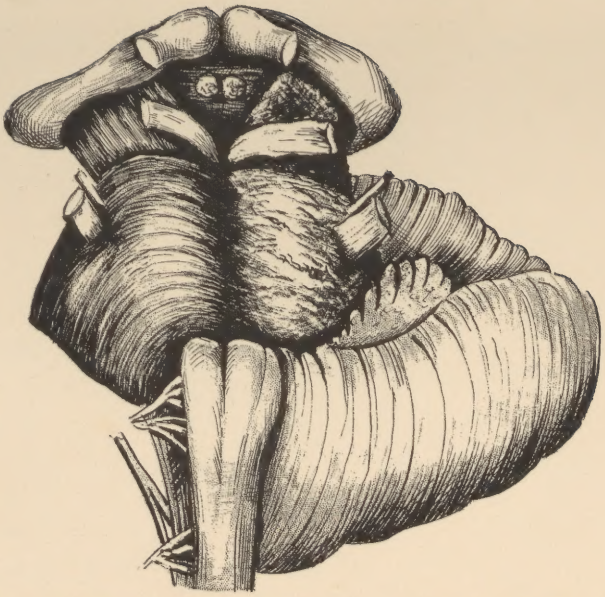


FIG. 1.—Brain axis, showing atrophy of the left crus and the left half of pons.

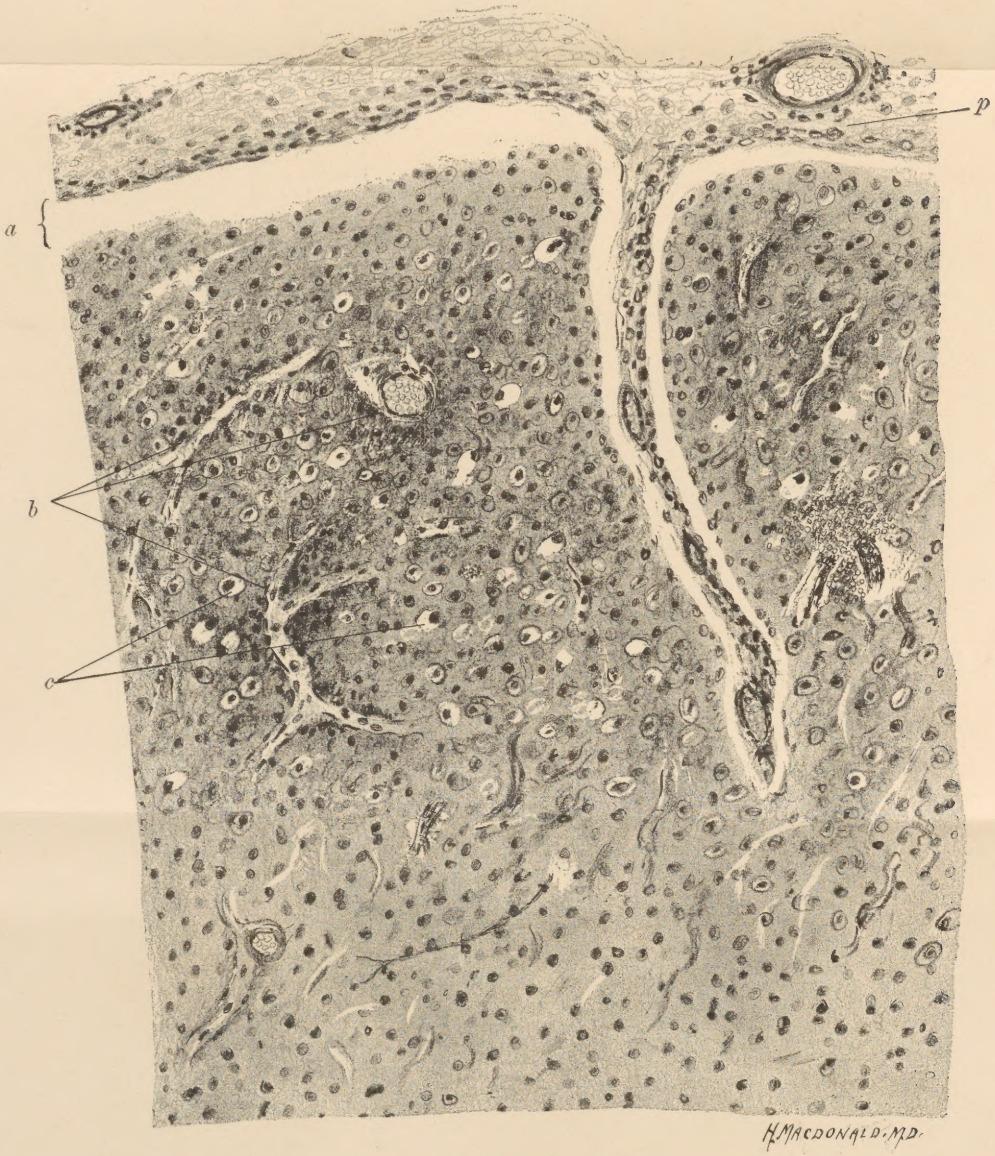


FIG. 2.—Drawn from microscopical section through the motor cortex, in a case of spastic paraplegia. *a*, Demarkation between cortex and pia. *b*, Blood-vessels in transverse and longitudinal section, showing infiltration of their walls. *c*, Altered cells with pericellular spaces. *p*, The pia, thickened and infiltrated, sending projection downward between two convolutions.

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